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Evan Syndrome: A Case Report

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ABSTRACT

Evan syndrome is an autoimmune disorder in which auto-antibodies are directed against antigens specific to red blood cells, platelets, and neutrophils. Also known as Immune-mediated thrombocytopenia [ITP], Autoimmune hemolytic anemia [AIHA], or immune pancytopenia. Clinical symptoms are thrombocytopenia, hemolysis, and severe anemia. The first line treatment for Evan syndrome intravenous corticosteroids or intravenous immunoglobulins and the second line treatment with rituximab or mycophenolate mofetil or splenectomy for those who are refractory to steroids. We report the case of a 35-year-old female who presented with high-grade fever with chills and rigors associated with hematuria and she has undergone a diagnosis of Evan Syndrome.

KEYWORDS: *thrombocytopenia, immune pancytopenia, immunoglobulins*

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INTRODUCTION

Evan syndrome is an autoimmune disorder in which auto-antibodies are directed against antigens specific to red blood cells, platelets, and neutrophils. Evans syndrome is characterized by warm AIHA, as opposed to cold AIHA, in which IgG antibodies react with antigens on the surface of red blood cells (RBCs) at body temperature. The immune system targets platelets carrying GPIIb/IIIa in ITP. Recently, a proposition has been laid out to classify the condition as primary (idiopathic) or secondary (associated with an underlying disorder) [1]. Diseases include autoimmune lymphoproliferative syndrome (ALPS), common variable immunodeficiency (CVID), and systemic lupus erythematosus (SLE), Leukemia chronic cystic (CLL) has been linked to secondary Evans syndrome [2].

Case Description

We report the case of a 35-year-old female admitted to the Intensive Care Unit of Saveetha Medical College and Hospital with complaints of high-grade fever with chills and rigors associated with hematuria from the past 2 days. She is a known case of Immune-mediated thrombocytopenia [ITP], AIHA, and hypothyroidism and she was on medication tablet thyronorm 100mcg, tablet wysolone 10mg.

At the presentation, she was looking dull, conscious, and oriented to time, place, and the patient with a Glasgow Coma Scale [GCS] 15/15. There are no other significant findings on general, physical, and systemic examination. Her vitals were stable.

All of the routine biochemical, microbiological, and serological tests were done which were normal. Her hematology values were a decreased hemoglobin value of 5.8, a Red blood cell count of 2.5m/mcl, platelet count of 25,000. Peripheral blood smear report showed normocytic anemia with neutrophilic leucocytosis. The direct Coombs test was positive.

Initial treatment at the time of admission 2 units of packed red blood cells were transfused and she was monitored and administered antibiotics and Vitamin B12, Paracetamol1g. Her blood parameters showed some improvement, although she hadn't reached the normal range, her health was steadily improving both clinically and hematologically while receiving intravenous steroids. On days 3, 4, and 5, she received further transfusions of 1 unit of PRBC and 1 unit of single donor platelet. Intravenous immunoglobulin [IVIG], Rituximab, a second antibody, was started, and the steroid dosage was

reduced. Her vital signs, blood indices, and fluid intake and output were monitored.

The outcome of the above treatment came to be good. Her condition gradually improved, a direct Coombs test showed negative and she was discharged from the hospital on the 8th day and visits the doctor once a week. No further complaints were received during her initial follow-up.

Discussion

We reported a case of 35-year-old female affected by Evan Syndrome she has already been diagnosed with immune-mediated thrombocytopenia and AIHA. The most prevalent antigenic targets in ITP are GPIb and Platelet Surface GPIIb/IIIa [3]. The cytometric examination revealed no increase in the number of B cells expressing GPIIb/IIIa antibody, indicating that autoantibody adhering to GPIb on the platelets was the cause of thrombocytopenia. These findings suggest that anemia in our patient is partially caused by immunological alteration as well as thrombocytopenia.

The AIHA is characterized by shortened red blood cell survival and a positive Coombs test. The responsible autoantibodies may be either warm-reactive or cold-reactive. The rate of hemolysis and the severity of the anemia may vary from mild to severe and life-threatening [4].

Hemolysis in warm antibody AIHA, which accounts for 80–90% of adult cases, is caused by antibodies that attach to red blood cells at 37 °C (98.6 °F). In our patient anemia improved with PRBC, single donor platelet transfusions, and IVIG, corticosteroids. There have only been two instances that we are aware of that IVIG or corticosteroids are effective.

These findings suggest that anemia in our patient is partially caused by immunological alteration as well as thrombocytopenia. In situations when laboratory results indicate a severely low platelet count along with a low hemoglobin level, Evans syndrome should be retained in the differential diagnosis list. Evans syndrome is diagnosed using a range of specialized testing, a comprehensive clinical assessment, a complete patient history, and the identification of specific symptoms. Evans syndrome cannot be definitively diagnosed with a single test; instead, a diagnosis is determined only after ruling out other probable causes. Evans syndrome is specifically diagnosed in patients who have both autoimmune hemolytic anemia (with a positive direct Coombs test) and ITP, even if they do not present simultaneously [5].

Intravenous immunoglobulin is given as a life-saving measure in situations of severe immune thrombocytopenic purpura presentations, and steroids such as prednisone and prednisolone are the first-line treatment. For refractory Evans syndrome, rituximab, sirolimus, cyclosporine, vincristine, azathioprine, mofetil mycophenolate, and thrombopoietin receptor agonists are among the second-line treatments. The woman in our case also underwent platelet and blood transfusions, responded favorably to rituximab, and had corticosteroids; our course of therapy was consistent with the approaches described in previous publications [6]. After she briefly became steroid-refractory, we moved from intravenous steroids to antibody rituximab, and as of right now, she is responding favorably to weekly steroid pulse treatment. Steroids function by eliminating macrophages that degrade an individual's red blood cells and platelets. Although concentrated immunoglobulin G derived from human plasma donors inhibits the FCy receptor of the macrophages' it remains a controversial therapy. Hematopoietic stem cell transplantation has been successful in patients who have not responded to immunosuppressive medications, however, it is important to take into account any potential major side effects [6].

Conclusion

All cases of ITP and AIHA should be thoroughly investigated to diagnose associated with other syndromes and to determine further treatment. In the case of Evan Syndrome, the patient responds to prophylactic treatment. She was discharged from the hospital on the 8th day and advised to visit the doctor once a week.

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